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BY

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A CASE OF PARTIAL EPILEPSY, APPARENTLY DUE TO A LESION OF ONE OF THE VASO- MOTOR CENTRES OF THE BRAIN.

ADA BROOK, æt. $6\frac{1}{2}$. Both father and mother are living and healthy; both are steady; there is no blood-relationship between them. Two other children are living and healthy; three are dead, one by an accident, and the other two, who were twins, in early infancy. The mother never miscarried. The father had an uncle who is said to have died 'out of his mind;' he was not, however, in an asylum, but died in his own house. There is no other history of insanity, and no history of fits in the family. Several members of the father's family are said to have died of consumption.

The patient was born with a very extensive 'mother's mark' on the right side of the head and face. The mark is bounded pretty accurately by the middle line in the upper lip, nose, forehead, scalp, and back of the neck, extending a little beyond the middle line on the chin and on the upper part of the sternum. It extends as low as the third or fourth dorsal vertebra behind, and the second costal cartilage in front. The lips, gums, tongue, roof of mouth, floor of mouth, uvula, and pharynx are all similarly affected, to a greater or less extent, on the right side. There is in addition a patch about the size of the palm of the hand over the left eye, frontal, and temporal regions. The mark is everywhere of a deep purple colour, the colour partially disappearing on firm pressure. All the parts affected are distinctly larger than the corresponding parts on the opposite side.

The right eye is affected in a similar way both in its superficial and deep structures. It is larger than the left;

the sclerotic is more vascular than usual, and with the ophthalmoscope it can be seen that the retina and choroid are involved. Mr. Nettleship has kindly examined the patient for me, and he has furnished me with the following most interesting and valuable report:—

‘*Measurements relating to the two Eyes (only approximate, as she is too restless for anything better).*

‘*Palpebral fissure.*—*Right*, 25 mm.; *left*, 22 or 23 mm.

‘*Cornea* (trans. diam.).—*Right*, 12 or 13 mm.; *left*, 10 mm.

‘*Pupil* (trans. diam.).—*Right*, 5 mm.; *left*, 4 mm. Pupils equally active to light.

‘*After atropine.*—The right pupil is rather oval in vertical direction, and more excentric inwards than usual (but atropine may not have acted fully).

‘*Vascularity.*—Visible vessels of right eye all larger than those of left; whilst downwards and inwards in front part of ciliary region of right eye is an ill-defined purplish or lilac patch seated in the sclerotic (scleral nævus).

Sight.—Cannot be accurately tested.

‘*Refraction* (by ophthalmoscope).—*Right*, myopia about $\frac{1}{10}$; *left*, hypermetropia at least $\frac{1}{4}$, and probably more.

‘This, therefore, proves the right eye to be really longer than the left, unless the myopia be due to increased convexity of the lens or the cornea; the former is unlikely, and of the latter there is no evidence. The globe is probably too large in all directions, and therefore too long. There is no myopic crescent, i.e., no evidence of bulging localised at the posterior pole of the eye, as is so very common in ordinary myopia.

‘*Ophthalmoscope.*—Right disc much redder than left; some of the right retinal veins are very tortuous, chiefly some of the large upward subdivisions: the tortuosity affects certain large branches or parts, and not the whole of any primary division; nor does it go so far as the disc. In the inverted image all the parts look smaller in the right than in the left eye, because of the difference of refraction.

‘*Choroid.*—The general colour is very markedly darker and at the same time redder in the right than the left eye. The difference is not what we should expect if it were due to the pigment being more abundant in one eye than the other; it is suggestive of venous or venous and capillary hypertrophy like that on the skin.’

After her birth the child enjoyed good health till she was six months old. She then began to have attacks of twitching in her left side, affecting the face, arm, and leg. The mother describes the fits as lasting for ten or twelve minutes, and consisting of little jerks which occurred every two or three seconds during that period. She had several of these attacks every day. As time went on the attacks became stronger, but the child did not seem to lose consciousness. When she got older her mother found that after the attacks she was weak in the left side, and could not walk so well as at other times. The fits continued to get stronger, the shaking of the left side was more severe, and after a time the right side began to be affected also. About eighteen months or two years ago she began to lose consciousness, and now the fits did not recur so often. The twitchings which occurred without loss of consciousness disappeared, and their place was taken by severe fits in which consciousness was lost, and there was very marked convulsion. These fits only occurred about once in every three, four, or six months.

Her mother describes the fits as she now has them as follows:—She first complains of a feeling in the palm of the left hand; the feeling appears to be a sort of painful tingling. She tells her mother a fit is coming on. The sensation in the hand lasts for several minutes, and she then clenches the hand firmly and bends up the arm. Sometimes both hands are clenched. A few seconds afterwards she falls down unconscious, but is not much more convulsed. After the fit is over the child sleeps soundly for some hours.

Sometimes the child tells her mother that she has a fit, but without any outward signs being seen. When asked what she feels, she says that it is a 'funny feeling' in the left hand.

She is troublesome and very restless.

Under the influence of bromide of potassium the patient has considerably improved. She has had fewer fits, and has been less restless.

Remarks.—I have no intention of going into the difficult question of the pathology of port-wine mark. The point to which I wish to call particular attention is the probable relationship between the mark and the fits. It will have been observed that the fits were for a long time very partial, being confined to the left side only, and apparently unattended by loss of consciousness. Now fits of this kind are rare in con-

nection with very young children, and I shall be able to show their significance more easily by saying a few words on the nature of epilepsy. It is becoming more and more certain that the phenomena of an epileptic fit or of an infantile convulsion are due to a liberation of force in an irregular and un-governed manner in the nerve-cells of some part of the grey matter of the cerebro-spinal system. This irregular liberation of force is very well described under the term, now so commonly used, of 'nerve-discharge.' A nerve-discharge which gives rise to convulsions is evidently due to some instability in those groups of cells where it takes place. This instability may be due to some inherent fault of construction (so to speak) of the grey matter, whether from a hereditary tendency to badly-formed nerve-tissues, or as the result of bad conditions in the parent at the times of conception and development, or from other causes. In such a case the tendency is for large tracts of grey matter to be affected in the same way, and for the two sides of the brain to resemble one another more or less in this respect. Hence in these cases of idiopathic epilepsy we usually meet with the severer forms of an epileptic fit, where there is instantaneous loss of consciousness, with little or no warning, accompanied by rigid contraction of nearly every muscle in the body. Sometimes, however, even in cases of widespread tendency to instability of grey matter, some small tract may be more unstable than the rest. In such a case it alone discharges at the beginning of the fit, producing the phenomenon of the warning, which may be mental, sensory, or motor, according to the seat of the discharging tract. Nerve-cells, however, intercommunicate freely with one another by connecting fibres, and when one cell is in action it tends to excite a similar state of activity in those with which it is brought into connection. It can easily be understood, then, that a disturbance such as that which is produced by a sudden and irregular discharge in a group of cells, propagates itself rapidly to neighbouring tracts. These being themselves over-excitabile, though to a less extent than those cells where the discharge has begun, an irregular discharge takes place in them, producing the phenomenon of the fit following the warning.

Epileptic fits may, however, be due not only to an idiopathic morbid excitability of grey matter, but grey matter which is in itself quite healthy may be induced to discharge irregularly under the influence of external irrita-

tion, such as results, for instance, from the presence of tumours, of masses of fibrous tissue growing down into the cortical substance from the meninges, after attacks of sub-acute or chronic meningitis; or again, from increased blood supply, as in acute meningitis. In such cases the discharge of nerve-force is very commonly confined at first to a small area. If the exciting cause be strictly localised, as in the case of a cerebral tumour, this area will be that which is in immediate contact with the tumour, but if the cause be more uniformly diffused, as in vascular disturbance, the discharge will begin in any part which is normally a little less stable than the rest; for doubtless in many healthy, as well as in many epileptic brains, there are regions which are more prone to take on irregular action than others. But a sudden, strong, and irregular discharge taking place in certain cells, must act as a strong excitant to other cells which are in communication with them. At first the passage of this stimulus is to some extent resisted by the intercommunicating fibres which are unused to their new function. We all know how much more easily nerve-stimuli are conducted after frequent repetition than on a first trial. The cells, however, which border the discharging area are as yet healthy, have no tendency to discharge, and are able to resist the rude shocks to which they are subjected. After a certain number of repetitions of the shocks, these cells begin to give way and finally become involved in the nerve-discharge. Hence we frequently find that in the early stages of tumours of the brain where fits are present, these are slight and confined to a very limited part of the body, as, for instance, one side of the face and certain muscles of one arm; and at this stage they are often unaccompanied by loss of consciousness. If consciousness be lost, it is usually after a very prolonged warning which the patient feels to be creeping gradually from one part of his body to another, or gradually passing from mere sensory phenomena to muscular action. As time goes on, however, the fits extend over more ground, and at the same time the warning becomes shorter; that is to say, after every new morbid discharge, not only do the nerve-fibres become more prone to conduct the stimulus, but the internal economy (so to speak) of the nerve-cells is to some extent upset, their condition is interfered with, and they are predisposed to discharge again more easily than before. Hence it is no uncommon thing for patients to say, 'My fits used only to be on one side, but now they are on both;' or still more often, 'I used to know

when my fits were coming on, but latterly I have had no warning;' and a friend who sees the fits will often tell us, 'The fits always begin on one side, but after a minute or two the patient struggles on both sides.'

Now everyone knows that, *cæteris paribus*, the grey matter is less stable in children than in adults; where an adult has a rigor, a child has a convulsion. From this it results that it is much rarer for fits to begin slowly in a young child than in an adult, especially where the fits are due to a hereditary instability of nerve-tissue. Whenever in a child partial fits occur, or fits preceded by a considerable warning, they may be taken as indicative of some external morbid influence acting on grey matter which is naturally healthy. In the patient I have shown, the fits for a long time were confined to the left side opposite to the port-wine mark; and the mother says that there appeared to be no loss of consciousness until she was three or four years old. They then involved the right side as well, and had become more severe. Even now, however, they begin in the left hand with a sensory warning of considerable duration, and sometimes appear to consist only of a sensation of some kind in that hand. From the nature of the fits, and from their mode of onset, I think there can be no doubt that they are due to some cause external to the nerve-tissue, rather than to an inherent instability of grey matter, and this external cause is in all probability to be found in the presence of a 'port-wine mark' on the surface of the right side of the brain, just as we have found it in the skin, mucous membranes, and retina of that side.

